Paediatrics Among Ethnic Minorities

Families from the Mediterranean and Aegean

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Greek Cypriots and Greeks

The groups of major importance, roughly in order of the size of their communities in the United Kingdom are: Greek Cypriots, Turkish Cypriots, Greeks, Italians, and Turkish families from the mainland.

Both Greek Cypriots and Greeks have been in Britain for many years, mainly in our large cities. Greek Cypriots originally came to London in small numbers in the 1920s, but much larger immigrations occurred in 1950-60 and again in 1974. In London the Greek community originally settled in Soho and the Cypriots in Camden Town. The main areas in London where Greek Cypriots now live are: Camden, Hackney, Haringey, and Islington; Greek families are mainly in north London. There are Greek and Greek Cypriot communities in most of the larger cities in Britain. Members of both communities are largely employed in the clothing trade, in restaurants and groceries, and in other service industries.

Religion—The Greek Orthodox Church, to which both communities belong, has not raised any moral or ethical difficulties about fetal diagnosis or termination of pregnancy in relation to β thalassaemia (for further discussion on genetically determined diseases see below).

Language—Greek is usually spoken at home, with English as the second language. There are rarely serious language difficulties at entry to school.

Naming system—In Greek Cypriots the wife takes the husband's surname, and the children take the father's first name as their surname. In Britain, however, children increasingly take their father's surname, thus conforming to British practice. Greek women may retain their maiden name after marriage or put their unmarried surname before their husband's surname; the children may use their mother's "surname" as theirs or use their father's surname. With Cypriot families it is most important to find out whether the family comes from the Turkish or the Greek part of Cyprus. Some idea may be obtained from the religion recorded in their notes: Greek Cypriots may be recorded as Christian, or as belonging to the Greek Orthodox church, while Turkish Cypriots are Moslems. Some indication may be obtained from the names. Common Greek or Greek Cypriot boys' names are: Antonis, Christos, Petros, Savvas and girls names: Eleni, Maria, Niki. Many Greek or Greek Cypriot surnames end in *ou* (Georgiou), meaning "son of" or in *aides* (Nicolaidis).

GENETICALLY DETERMINED DISEASES

β Thalassaemia (*Mediterranean anaemia*, *Cooley's anaemia*)

β Thalassaemia, inherited as an autosomal recessive disease, occurs in both communities but is more common among the

Cypriots. In the homozygous state (thalassaemia major) there is an inability to make adult haemoglobin (HbA). Heterozygotes (thalassaemia minor, thalassaemia trait) are invariably asymptomatic but can be reliably detected by screening. In north London Modell has shown that about 17% of the Cypriot (Greek and Turkish) community have thalassaemia minor and 3% of marriages have a one in four risk of producing a child with thalassaemia

World distribution of β thalassaemia. Reproduced from *Thalassaemia Syndromes*. 3rd ed. Weatherall DJ, Clegg JB. Oxford: Blackwell Scientific Publications, 1981, with permission of authors and publishers.

major¹; without intervention about one in every 120 live births within this community would be expected to be of an affected child.

Clinically, thalassaemia major usually presents between the ages of 6 months and 1 year with pallor, listlessness, and failure to thrive; severe anaemia occurs, often with a haemoglobin concentration as low as 3.5 g/dl, and abdominal distension due to hepatosplenomegaly. Older children, if untreated or undertreated, may develop the so called "Mongolian" facial appearance due to expansion of the frontal and maxillary bones. Blood films show red cells of irregular shape and size, with numerous target cells. On electrophoresis there is a high concentration of fetal haemoglobin (HbF) and an almost complete absence of HbA; concentrations of HbA₂ are normal or slightly raised and are of no diagnostic importance. Children with suspected thalassaemia major should be admitted to hospital immediately for confirmation of the diagnosis and transfusion. They should be looked after at a special clinic or by a paediatrician or haematologist with a particular interest in the condition. Treatment consists of regular blood transfusions and

injections or infusions with desferrioxamine to reduce the overload of iron.

Though thalassaemia minor is of no clinical importance, its detection is important in preventing thalassaemia major. The characteristic microcytosis of thalassaemia minor can be detected by a Coulter counter and confirmed by a high concentration of HbA₂. With the support of a well informed and enthusiastic community the incidence of thalassaemia major in north London has been reduced by 60% over the past seven years. All Greek and Cypriot women (and also in practice all immigrant women are included) are screened at their first attendance at the antenatal clinic; the husbands of women with thalassaemia minor are then also tested. Couples who are both heterozygous are offered fetal diagnosis at 18 weeks of pregnancy; if the fetus is found to be homozygous termination of pregnancy is offered at 20 weeks or as soon after this as possible. Termination at this late stage is emotionally distressing and allows little time for discussion. Earlier fetal diagnosis, at 8-11 weeks, is now possible in a proportion of Cypriot families at risk, and as newer techniques come into use early diagnosis should be available to all Cypriot couples who need it. Obviously, with a community that is well informed about thalassaemia screening should be extended to school leavers, but this would need to be supported by a programme of education on the implications of having thalassaemia minor.

The family doctor and β thalassaemia—The possibility of thalassaemia major should be considered in young children with severe anaemia whose families have originated from Mediterranean or Aegean countries or Bulgaria, Soviet Central Asia,² or the Indian subcontinent and South East Asia. The figure shows the world distribution of β thalassaemia. Treatment of a child with thalassaemia creates an emotional and often economic strain on the family, who require support from their family doctor and the community services. Treatment with iron or other haematinics should not be given unless requested by a specialist clinic; folic acid is not required if the child is on a programme of regular transfusions. Parents or adolescents requesting screening, and the families of all patients with thalassaemia major and minor, should be referred to the nearest appropriate haematology department (see box for centres in London). Families or individuals requiring further advice should be referred to the United Kingdom Thalassaemia Society (see box).

Glucose-6-phosphate dehydrogenase (G6PD) deficiency

G6PD deficiency is transmitted through an x linked intermediate gene with variable expression in the heterozygous female (see below); the affected person has a notable reduction of the enzyme G6PD in the red cells, causing rapid haemolysis in response to several drugs (table I) and also during acute infections, particularly hepatitis, and in diabetic ketoacidosis. Neonatal jaundice is also a common problem. Three main variants of this condition occur: the Mediterranean, the African, and the Oriental forms. In the Mediterranean and Oriental forms, but not the African, contact with the broad bean (*Vicia faba*) produces a particularly severe haemolysis (favism); even sniffing the pollen may be enough to cause a haemolytic crisis. Table I summarises the main differences between the three types of G6PD deficiency.

TABLE I—The variants of G6PD deficiency

	Mediterranean and Oriental*	African (including West Indians)
Enzyme activity as normal (%)	0-5%	5-15%
Latent period after exposure (hours)	3-24	24-36
Haemoglobinuria	Always	Common
Anaemia	Haemoglobin often <6 g/dl	Haemoglobin rarely <6 g/dl
Duration of haemolysis	Continuous if drug is continued sometimes fatal	Self limiting even if drug is continued (not advised); rarely fatal
Favism	Yes	No
Neonatal hyperbilirubinaemia	Common, without drug contact	Without drug contact, in preterm and term infants
Affected females	Common	Common

*Though genetically distinct the Mediterranean and Oriental variants behave in a similar manner.

Useful addresses

United Kingdom Thalassaemia Society, 107 Nightingale Lane, London N8 (01 384 0437)

Cypriot (Greek) Advisory Service, 26 Crowndale Road, London NW1 1TT (01 388 7971/2)

Cyprus Turkish Association, 34/35 D'Arbly Street, London W1A 4YL (01 437 4940)

For blood tests for thalassaemia contact via general practitioner or direct:

North Middlesex Hospital, Sterling Way, London N18 (01 807 3071, ext 471)

Department of Haematology, Royal Free Hospital, Pond Street, London NW3 (01 794 6500)

Dr B Wonke, Department of Haematology, Royal Northern Hospital, Holloway Road, London N7 6LD (01 272 7777)

Department of Haematology, University College Hospital, Gower Street, London WC1 (01 387 9300)

TABLE II—Drugs likely to cause haemolysis in G6PD deficiency

Antimalarials	Primaquine Pamaquin Pentaquine
Sulphonamides	Sulphanilamide Sulphacetamide Sulphapyridine Sulphamethoxazole (in Bactrim and Septrin preparations) Silver sulphadiazine (surface application for burns)
Sulphones	Dapsone (in Maloprim preparation, prophylaxis for malaria)
Nitrofurans	Thiazosulphone Nitrofurantoin (Furadantin preparation)
Analgesics	Acetanilide
Miscellaneous	Methylene blue Nalidixic acid (Negram preparation) Naphthalene Niridazole Phenylhydrazine Toluidine blue Trinitrotoluene

Table (modified) from: Wintrobe MM, Lee GR, Boggs DR, et al. *Clinical Hematology*. 8th ed. Philadelphia: Lea and Febiger, 1981:992, with permission of authors and publishers.

Haemolytic crises—As would be expected males are more often affected, but in all three types the gene is so common that homozygous females, having received the gene on the X chromosome from each parent, are not uncommon. Homozygous females are usually as severely affected as males, while heterozygous females may be mildly affected. The haemolysis is so rapid that severe anaemia, often down to a haemoglobin concentration of 3-4 g/dl, develops before any jaundice becomes obvious. Haemoglobin should be looked for in the urine as evidence of haemolysis. The patient requires immediate admission to hospital for transfusion. Confirmation of the diagnosis may have to await the disappearance of transfused cells and the maturation of the young cells produced by the patient in response to haemolysis. The diagnosis can, however, usually be inferred by investigating the parents and siblings.

Neonatal jaundice—In some enzyme deficient infants neonatal hyperbilirubinaemia occurs without any drug contact. Jaundice usually develops on the second or third day, reaching a peak around the fourth to fifth day. Exchange transfusion may be required, particularly in preterm infants or when there are other factors causing hyperbilirubinaemia. The possibility of G6PD deficiency should be considered in infants of either sex with immigrant parents or mothers (G6PD deficiency, apparently of the Mediterranean type, is not uncommon in the Indian subcontinent). It is rare for parents to be aware of the disease in their family. When the diagnosis has been confirmed in the child the rest of the family should be investigated and the clinical and

genetic implications fully explained. Affected people and their family doctor should be given a list of the drugs that must be avoided (table II).

Turkish and Turkish Cypriot families

A few Turkish Cypriots came to Britain in the 1920s, but many more arrived in 1947-50 and again in 1960-2. In London they have settled mainly in the north (Haringey, Islington, Stoke Newington), the east (Hackney), and the south east (Bromley, Catford, Lewisham, New Cross). There are smaller communities in most of the larger cities. Turkish families came to Britain more recently and have tended to settle in the same areas as the Cypriots. Members of both communities work in the clothing industry, in groceries and restaurants, and in other service industries.

Religion—Both communities are Moslem, though with usually less strict observance than in immigrants from the Middle East and Pakistan or Bangladesh. There is an absolute prohibition on eating pork or pork products but no insistence on halal meat. Marriage is monogamous (by law in Turkey). Problems about necropsies are rare, but the parents should be approached with gentleness and tact. Parents should also be consulted about preference for a woman doctor if their daughter has to be examined at the surgery or outpatient clinic.

Language—Turkish is spoken at home by both groups, but Turkish parents may have no second language, except possibly German if they have worked in West Germany. Learning English presents difficulties for Turkish adults, and an interpreter may be required. Pronunciation of Turkish names is not as difficult as it may appear, as it is largely phonetic: ç is pronounced "ch" as in church; ş is "sh" as in shoe; ö and ü are as in the German; ı is the English e as in meat; i is as "oer"; and ğ is soft with a prolonged preceding vowel.

Naming system—The naming system is usually the same for Turkish and Turkish Cypriot families, but it is impossible to generalise. The wife and children usually take the father's first name as their surname. Many families, however, have adopted the British system and take the father's family name (surname) as their own. It is essential to find out which system the family is using. Typical boys' names are: Gültekin, Mehmet, Suleyman, Turgay, Yusuf and girls' names: Ayşe, Gulay, Hatije, or they may be the same as a boy's name but with a terminal e as in Ermin(e). The ending *oğlu* means "son of."

Medical conditions found in the children—β Thalassaemia and

G6PD deficiency occur in both groups with a higher incidence in Turkish Cypriots, though it is probably not as high as that in Greek Cypriots. There are no other conditions specific to these two communities.

Italian families

Italian families, mainly in the catering trade, have been in Britain for many generations; some immigration occurred in the Luton and Bedford areas after the end of the second world war, and there has been further immigration into larger cities since then.

Religion—Most Italians are Roman Catholics with a strong religious objection to termination of pregnancy, so fetal diagnosis is pointless. Nevertheless, and particularly in relation to β thalassaemia, it should not be assumed without discussion that all parents will have these views.

Language—Italian is normally spoken at home with English as a second language. There are rarely any serious language difficulties at entry to school.

Medical conditions found in the children—Both β thalassaemia and G6PD deficiency occur quite commonly, particularly in families originating from southern Italy, Sardinia, and Sicily.

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- 2 Weatherall DJ, Clegg JB. *Thalassaemia syndromes*. 3rd ed. Oxford: Blackwell Scientific Publications, 1981:299-300.

Further reading

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- Oski F, Naiman JL. *Hematologic problems in the newborn*. 3rd ed. Philadelphia: WB Saunders, 1982:112-8 (for G6PD deficiency and neonatal jaundice).
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(This article is the fifth in a series)

What is evoked response audiometry?

Evoked response audiometry is a commonly used term for the measurement of acoustically evoked electric potentials that assess the functional integrity of the auditory system recorded as an audiogram. Fifteen acoustically evoked potentials are known, but those in clinical use are cortical evoked potentials (cortical electric response audiometry), brain stem electrical potentials, and electrocochleography. The former are non-invasive in that the electrodes are placed on the vertex of the head and mastoid in the first, and on the forehead and ipsilateral mastoid in the second. The recorded electric potentials (such as electroencephalographic waves) in response to sound are so small that the response to repeated signals is summated and averaged by an electronic device (perhaps a computer) and displayed. With this method the minimal response to sound at different frequencies and intensities may be measured and an approximate pure tone threshold audiogram constructed.

The tests are objective and valuable in assessing the threshold of hearing in a deaf child and, most commonly, in an adult who exaggerates his hearing loss, usually for compensatory gain in medicolegal cases. In common jargon it is used as a "lie detector," for it measures the cortical responses to sound objectively by electroencephalographic methods. In electrocochleography the cochlear activity "or the level of the sensorineural loss" is recorded by a needle electrode pushed through the tympanic membrane to lie on the medial wall of the middle ear. Although more accurate, it is less convenient as minimal surgery is needed to place the electrode, and a surgeon is

required, whereas the cortical and brain stem electric potentials acoustically evoked can be done by trained technicians.—J D K DAWES, reader in otorhinolaryngology, Newcastle upon Tyne.

What is the drug (if any) in the plant feverfew (*Pyrethrum parthenium*)? What are its therapeutic benefits?

Feverfew—now classified as *Tanacetum parthenium* Bernh—is used extensively as a herbal treatment for migraine, arthritic conditions, psoriasis, etc. The average daily intake seems to be equivalent to about 100 mg of the dried leaf. Aqueous extracts inhibit the biosynthesis of prostaglandins at two points—the phospholipase mediated formation of arachidonic acid from platelet lipids, and the subsequent conversion of arachidonic acid to prostaglandins by the prostaglandin synthase system. Most of the drugs used in migraine and arthritis are prostaglandin inhibitors, so the efficacy claimed for feverfew in these conditions may be justified on this basis.¹ Feverfew contains the germacranolide, parthenolide, which has been shown to be a prostaglandin inhibitor (Pugh WJ, Sewell RDE, in preparation) and also possesses cytotoxic properties against normal human fibroblasts and human laryngeal carcinoma cells.²—W J PUGH, lecturer in pharmaceutical chemistry, Cardiff.

- 1 Makheja AN, Bailey JM. The active principle in feverfew. *Lancet* 1981;ii:1054.
- 2 Lee K-H, Huang E-S, Piantadosi C, Pagano JS, Geissman TA. Cytotoxicity of sesquiterpene lactones. *Cancer Res* 1971;31:1649-54.